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Case Report

Lymphoma in Ovary – A Rare Case

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Abstract

However, Primary ovarian lymphoma (POL) is rare, accounting for 0.5% of all non-Hodgkin's lymphoma and 1.5% of all ovarian neoplasms. We present a case of ovarian non-Hodgkin's lymphoma, manifesting like an advanced ovarian cancer, which was managed by surgery and chemotherapy.

Keywords: Ovary; Lymphoma; Chemotherapy

Introduction

Involvement of the ovary by malignant lymphoma is well known as a late manifestation of disseminated disease. However, Primary ovarian lymphoma (POL) is rare, accounting for 0.5% of all non-Hodgkin's lymphoma and 1.5% of all ovarian neoplasms [1]. We present a case of ovarian non-Hodgkin's lymphoma, manifesting like an advanced ovarian cancer, which was managed by surgery and chemotherapy.

Case Report

A 45-year-old premenopausal woman (gravida 1, para 1) presented with history of pain in right side of lower abdomen, dull aching in character for 4 months which increased in severity progressively. It was associated with feeling of heaviness in lower abdomen for 2 months. She had no other systemic complains. She had normal and regular menstrual cycles with one live issue 17 year back. No other significant co-morbidities were present. On examination, she had a large abdominopelvic mass present which was firm to hard in consistency and arose from the adnexal region. CECT abdomen and pelvis showed a heterogeneous mass in lower abdomen with minimal ascites. CA-125 level was 85.93. Chest X-ray and thyroid profile was normal. She underwent Exploratory laparotomy with Total Abdominal Hysterectomy and Bilateral Salpingo-oophorectomy. Intra-operatively, there was a 20 × 15 cm size irregular mass, firm in consistency was seen arising from Right ovary. Final Histopathology reported it as Non-Hodgkin Lymphoma of right ovary. Morphologically it was classified as Diffuse Large Cell B Cell Lymphoma (DLBCL). On Immunohistochemistry, CD 20 and LCA was positive, while CK, PLAP and CD 3 were negative. With the histopathological diagnosis of Right ovarian NHL, patient underwent further staging evaluation. No systemic lymphadenopathy was found. CECT neck, chest, abdomen and pelvis were normal except for post hysterectomy status. Bone marrow aspiration and biopsy didn't showed infiltration. CA - 125 was 9.4 post operatively. Patient was staged as Ann Arbor Stage IE and was started on systemic Chemotherapy with R-CHOP regimen comprising Rituximab, Cyclophosphamide, Vincristine and Prednisolone in standard doses. Patient has received 6th cycle in February 2011 and is on regular follow-up with no clinical and radiological evidence of disease.

On 20/1/14 patient come in RT OPD with complains of weakness with intermittent loss of vision and hearing On systemic examination No pallor/jaundice/cyanosis /clubbing Vitals WNL No lymphadenopathy Chest- WNLCVS- WNL Abdomen - Soft and non tender No hepatosplenomegaly, no ascites WNL B/L decreased for both near and distant. Pt referred to ophthalmology and advice CEMRI brain. Ophthalmology examination Fundus examination-B/L severe disc edema with peripapillary hemorrhage with peri-

retinal bleeding. CEMRI: (21/1/14) shows widened subarachnoid space around B/L optic nerve subtle perineural heterogonous enhancement especially in retro bulbar part. Pt admitted in neurology ward Started on steroids and CSF cytology done CSF cytology (27/1/14) shows infiltration by lymphoma. Patient started on whole brain radiation from 29/1/14 30 Gy/10#/2 week, with intra thecal methotrexate bi weekly RT completed on 8/2/14. Total 6 cycle of methotrexate given last on 17/2/14 CSF cytology negative after 1st cycle. On last follow up on 17/2 /14 Pt symptomatically better no complains. Vision - normal in both eye

Discussion

Malignant lymphoma is revealed by an ovarian mass in less than 1% of all non-Hodgkin lymphoma, while autopsies performed on patients with non-Hodgkin lymphoma (NHL) show an ovarian enlargement in 7 - 26% [2]. Primary ovarian lymphoma (POL) is still much rarer and represents 0.5% of NHL and 1.5% of all ovarian neoplasms [1]. Diffuse lymphoma and a B-cell phenotype are the most common histological type and phenotype, respectively [1,3,4].

The distinction between primary and secondary lymphomas is usually made postoperatively, when the diagnosis of ovarian lymphoma is established and when its extension is evaluated. Fox., *et al.* [5] have proposed the stringent criteria for the diagnosis of a POL. Primary and secondary ovarian lymphomas differ in terms of prognosis. Contrary to POL, patients with an occult nodal lymphoma presenting as an ovarian mass have a poor outcome with 5 -year survival rate ranging from 7% to 38% [4,6,7].

Most patients of POL are young, with a median age of 42 - 47 years [1]. Fever, emaciation, or night sweats have to be investigated during the patient's interview. Clinical examination may reveal a palpable adenopathy, or a liver or spleen infiltration [1,7]. None of these clinical signs, except a relatively young age, was found in our patient.

Ferrozzi., *et al.* reported eight patients with ovarian NHL and assessed their most typical imaging patterns [8]. Ovarian lymphomas were frequently bilateral and homogeneous, without ascites, and the tumors always exceeded 5 cm in diameter. Ultrasonography showed homogeneous, hypoechoic, and mildly vascularized tumors. In all cases, computed tomography (CT) revealed clear-cut hypodense lesions, with mild contrast enhancement. In our patient, imaging was rather suggestive of an ovarian carcinoma. CT scan showed a heterogeneous mass, reshaped by several necrotic centers.

Lymphomas are treated with chemotherapy, using a regimen based on the cytological type of the NHL. There is not a standard treatment protocol for primary ovarian NHL because of its rarity. The best option seems to be chemotherapy according to the specific histology. A CHOP regimen (cyclophosphamide, Adriamycin, vincristine, prednisone), is usually administered.

Yamada., *et al.* report a case with a diffuse large B-cell ovarian malignant lymphoma, which presented as an advanced carcinoma successfully treated with eight IV CHOP regimens. A mixture of 100 mg of cisplatin, 400 mg of etoposide, and 1000 ml of saline solution was also administered in the peritoneal cavity because an undifferentiated carcinoma was suspected at the histological examination of the frozen section. The patient was alive and disease-free 6 years after the chemotherapy [6].

Niitsu., *et al.* obtained in a 54 -year-old woman the complete remission of a follicular lymphoma (grade 1, stage IIE) with a combination of cyclophosphamide, vincristin, bleomycin, etoposide, doxorubicin, and prednisolone (cycl OBEAP) [4].

Dimopoulos., *et al.* [1] reported complete remissions and long-term survivals in patients treated by combination chemotherapy appropriate to each histologic type. In all their patients, the diagnosis could only be established after a laparotomy, whereas the clinical findings suggested an epithelial ovarian neoplasm. Chemotherapy regimens included CHOP regimen or variants of CHOP. The authors obtained a complete remission in 64% of their patients. Long-term disease-free survival and overall survival were noted in 46% and 57%, respectively.

Conclusion

Our patient received chemotherapy with R-CHOP regimen for six courses and is clinically and radiologically disease-free. Primary ovarian lymphoma is rare and mimics other common gynecologic tumors. Outcome seems to be determined by its histologic type and phenotype, and by the chemotherapy regimen. Stage and mode of presentation also influence its prognosis.

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